

## Clinical Report 2 (Kampo Medicine)

*Cornelia de Lange Syndrome with Recurrent Pneumonia*

Hideaki Yamaguchi  
Tosei General Hospital

Case: 6-year-old girl with Cornelia de Lange Syndrome and recurrent pneumonia

The patient was clinically diagnosed with Cornelia de Lange Syndrome, and received six operations since her neonatal period. They included diaphragmatic hernia repair surgery, colectomy, colostomy, esophagocardioplasty, and pyloroplasty. She began to develop recurrent respiratory tract infections at the age of 1. At the age of 4, the frequency of her being hospitalized increased, and it eventually got to the point where she was admitted to the hospital eight times in a year, for a total of 100 days.

The patient showed no susceptibility to infection, and general examinations and immunological tests showed no abnormality. The recurrent pneumonia was thought to occur mainly from diaphragmatic paralysis on the left side. Kampo therapy was considered, as no effective control could be found in Western medicine.

Present status: Weight 11kg. No speech.

Treatment: Assuming a dual deficiency in the lung and spleen, qi and yin deficiency, and congestion of lung heat, the following prescription (decoction) was administered (daily dose; brewed and taken in 2 to 3 separate doses per day).

*Astragali* Radix 8g

*Ephedrae* Herba 1g

*Ophiopogonis* Radix 5g

*Armeniacae* Semen 3g

*Bupleuri* Radix 3g

*Platycodi* Radix 2g

*Ziziphi* Fructus 3g

*Glycyrrhizae* Radix 1g

*Gypsum* Fibrosum 4g (short-term usage)

Initially, the patient refused to take the prescription. However, her mother thought of ways of taking it, such as by baking a cake mixed with the decoction or adding it to the sauce when making simmered vegetables. As a result, the patient was able to take about two-thirds of the daily dose every day.

She was admitted to the hospital once after two-and-a-half months, but has not required hospitalization thereafter, and her symptoms markedly improved. The prescription was administered for a year and five months while making adjustments, and the symptoms have stabilized even after terminating the prescription.

Observation: Kampo therapy based on the reinforcing and reducing method is effective against recurrent infections. However, decoction is no less effective in flexibly addressing severe cases such as this particular case.

Reference: Cornelia de Lange Syndrome is a congenital disease that is characterized by delayed physical and intellectual development. Patients with Cornelia de Lange Syndrome have a distinct appearance, including a small head, thick eyebrows that meet in the middle, a small pointed nose, a small chin, excessive body hair, and small hands and feet. Cornelia de Lange Syndrome also accompanies various malformations of the organs and epilepsy. Certain genetic mutations are seen in roughly half of all patients.